SARCOIDOSIS VASCULITIS AND DIFFUSE LUNG DISEASES 2013; 30; 146-148

First presentation of sarcoidosis with severe obstructive sleep apnoea and epiglottic involvement

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Abstract. Sarcoidosis of the upper respiratory tract (SURT) is rare but well documented. We present a patient whose first presentation with sarcoidosis was with dysphonia, upper airways obstruction and severe obstructive sleep apnoea (OSA) . Systemic steroids and hydroxycholoroquine led to improvement in phonation, endoscopic appearance and upper airway obstruction with significant improvement in Apnoea Hypopnoea Index (AHI) on polysomnography. *(Sarcoidosis Vasc Diffuse Lung Dis 2013; 30: 146-148)*

Key words: Dysphonia, obstructive sleep apnoea, sarcoidosis

Abbreviation List

AHI: Apnoea Hypopnoea Index
OSA: Obstructive Sleep Apnoea
SURT: Sarcoidosis of the Upper Respiratory Tract
CPAP: Continuous Positive Airway Pressure

Introduction

Sarcoidosis is a multisystem disorder characterised by the presence of non caseating granulomas of unknown cause. It frequently presents with bilateral hilar lymphadenopathy, pulmonary infiltration, and ocular and skin lesions (1). Involvement of the upper respiratory tract is rare but well documented in the literature (2-4). We present a case report of a patient with sarcoidosis of the epiglottis as the first manifestation of the disease with resultant severe obstructive sleep apnoea. He was treated with systemic steroids and hydroxychloroquine with good response.

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Case Report

A 30 year old male ex smoker presented to the ENT service with a 4 week history of worsening dysphonia. He also complained of excessive snoring, daytime hypersomnolence and had witnessed nocturnal apnoeas. Cardiovascular and respiratory examinations were unremarkable. Chest radiography was normal. Laryngoscopy revealed supraglottic oedema and histology showed nonspecific inflammatory changes. The patient received a short course of oral corticosteroids but 3 months later presented with worsening symptoms. A repeat laryngoscopy and biopsy showed occasional small non-caseating granulomas. Two months later a further laryngoscopy showed thickened epiglottis, supraglottis and aryepiglottic folds. Biopsies demonstrated multiple non-caseating epithelioid granulomata within the subepithelial tissue suggestive of sarcoidosis. Cultures for mycobacteria and fungal pathogens were negative.

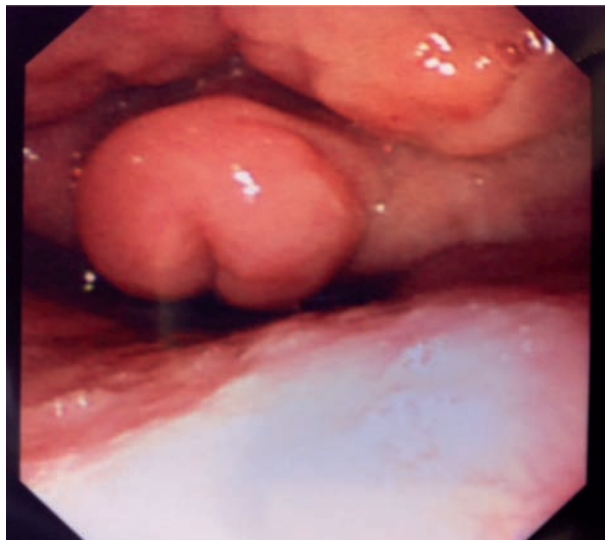

Fig. 1. Patient's epiglottis at first bronchoscopy

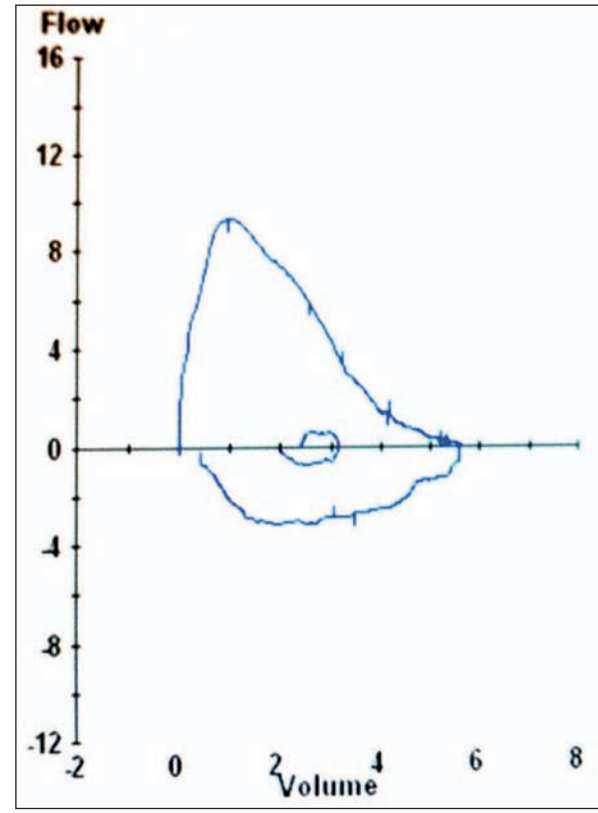

Fig. 2. Patient's flow volume curve prior to start of treatment, typical of extrathoracic compression

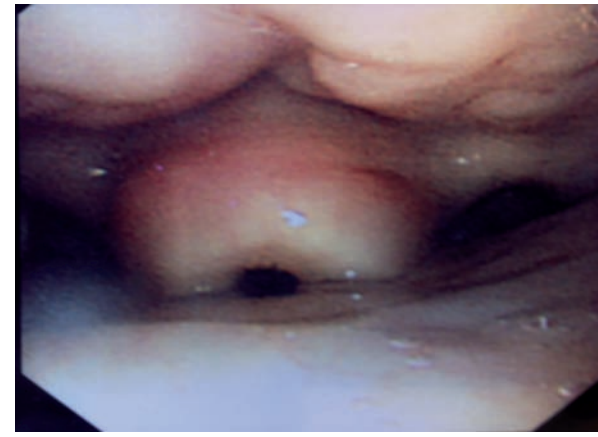

Fig. 3. Photograph of patient's epiglottis taken at bronchoscopy with reduction in swelling after corticosteroid treatment

The patient was referred to our respiratory service. On examination he had a BMI of 33 and neck circumference of 34 cm. He scored 10/24 on the Epworth sleepiness scale. Bronchoscopy revealed diffuse epiglottic enlargement (figure 1) and intubation was deemed unsafe. Flow volume study confirmed an attenuated inspiratory loop with reduced peak inspiratory flow of 3.23 L/sec typical of extra-thoracic obstruction (figure 2). Polysomnography confirmed severe OSA with an AHI of 43.2/hour. Computerised Tomography of the thorax showed areas of pulmonary opacification scattered throughout the right upper and bilateral lower lobes suggestive of sarcoidosis with superimposed aspiration. There were no enlarged mediastinal or hilar lymph nodes.

The patient was treated with high dose oral corticosteroids followed by hydroxychloroquine. Symptomatic and structural improvement was achieved confirmed by repeat polysomnography (with AHI falling to 5.4/hour), bronchoscopy (figure 3) and inspiratory flow volume loop. Currently he is on 10 mg prednisolone and 400 mg hydroxychloroquine once a day

Discussion

Laryngeal sarcoidosis is very rare. McCaffrey et al reviewed the records of 2319 patients attending the Mayo clinic with a diagnosis of sarcoidosis and found the incidence of laryngeal involvement to be less than 1% (4). Symptoms of laryngeal sarcoidosis include hoarseness, dyspnoea, dysphonia and, in severe cases, acute upper airway obstruction (5). Cur-

rently the only identifiable risk factor for laryngeal involvement in sarcoidosis is lupus pernio (6). In our literature search we identified only two previous case reports of significant obstructive sleep apnoea due to upper airway sarcoidosis (7, 8). In one of the two cases a diagnosis of sarcoidosis was pre-existing and in the second the disease was only involving the larynx. One study has estimated the incidence of OSA in sarcoidosis patients to be higher than controls (17% compared to 3%) (6). Usually this is related to long term steroid treatment, although neuromuscular involvement with sarcoidosis has been previously suggested as another mechanism (9).

Sarcoidosis and OSA are relatively common conditions. It is arguable that this patient had sleep apnoea and then developed concomitant sarcoidosis. The measurable improvement in polysomnography with immunosuppressive therapy supports our approach in treating this case as OSA secondary to laryngeal sarcoidosis (table 1). There was significant reduction in events even while supine confirming treatment response rather than other common causes of inter-night variability.

Our patient had no previous history of sarcoidosis and the lesion had to be biopsied 3 times before a definitive diagnosis was reached. This is not unusual. In one case series a patient had 3 biopsies positive for sarcoidosis out of 10 (10).

We opted to treat this patient with systemic steroids and then hydroxychloroquine with good symptomatic and measurable response. There are no randomised trials in the literature to guide treatment options of such cases. Antimalarial agents has been used previously for SURT with good results (11), although in general methotrexate has been used more commonly. Other options include intralesional steroids, Carbon Dioxide Laser resection (10, 12) or tracheostomy in treatment resistant cases or where acute airway obstruction is imminent (13).Whatever the choice is, we suggest that treatment should be initiated as soon as possible following the diagnosis of laryngeal sarcoidosis due to the critical location of the disease which can lead to severe airway compromise with deleterious effects.

Table 1. Polysmnographic parameters from baseline and follow up sleep studies

	Baseline polysomnography	Follow up Polysomnography
AHI	43.2	5.4
AHI supine position	48.3	8.9
AHI non supine position	38.9	12.7
RDI	43.2	11.5
Percentage of sleep time in supine position	45.9%	30.6%

RDI: Respiratory Disturbance Index

In their case report Shah et al described using CPAP as initial treatment for OSA in a laryngeal sarcoidosis patient with success followed by systemic steroids (7). We chose not to use CPAP and to treat with anti-inflammatory treatment. This approach proved successful. It remains to be determined if remission can be maintained on complete withdrawal of steroid treatment. In patients with sarcoidosis and OSA laryngeal involvement should be considered as a treatable cause of upper airway obstruction.

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